

CASE REPORT

Orbital Pseudotumor: Diagnosis and Management in A Secondary Hospital – A Case Report

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ABSTRACT

Objective: To increase the awareness in diagnosing orbital pseudotumor and its management, especially in a peripheral secondary hospital with limited facilities.

Case presentation: A 47 years old woman came with left eye (LE) protrusion, blurred vision, retrobulbar pain, and severe headache at regular intervals. This condition begun 8 months ago, and was diagnosed with orbital cellulitis, however the symptoms persisted until now. Physical examination of the LE showed a decreased in visual acuity, protrusion, chemosis and conjunctiva injection, increased in IOP, limited ocular motility, ipsilateral afferent pupillary defect and optic disc oedema. Plain and contrast CT-Scan showed a hypertrophy of ocular muscles with soft tissue swelling. Patient then diagnosed with idiopathic orbital inflammation and immediately prescribed with high dose steroid for 5 days with observation with glucose level observation for toxicity. Afterwards, she was given an oral corticosteroid and was tapered off for the next few weeks. At two months follow up, there were improvement in protrusion, motility and visual acuity.

Conclusion: A complete history taking, physical examination and appropriate additional examination are needed. In hospital or clinical setting with limited facilities, a thorough and comprehensive examination are needed in order to establish the diagnosis and management of orbital pseudotumor.

Keywords : Orbital Pseudotumor, Idiopathic Inflammatory Syndrome, Corticosteroid therapy

In 1905 Birch-Hirschfeld was the first one to found a term for Orbital Pseudotumor (OP) or also known as Idiopathic Orbital Inflammation (IOI), it is a benign disease with unknown etiology.^{1,2} Shields et al, showed that 135 of 1264 patients were diagnosed with pseudotumor orbita.³ Another study also showed that 10% of all orbital mass was counted for OP.^{3,4} This condition can affect both gender approximately equal, which was reported from Siriraj Hospital, that 49 patients, consist of 24 males and 25 females were diagnosed with OP and mean age was 43.75 years old.⁵

The clinical presentation can occur on acute, sub-acute or it could be develop to chronic phase and usually only affect one eye (unilateral), however occasionally it may become bilateral.⁶ The typical symptoms of OP are, orbital ache or headache, patients can also have proptosis, eyelid swelling, limited motility, tenderness to palpation, chemosis and visual loss.^{7,8} The additional examination includes computed tomography (CT) and/or magnetic resonance imaging (MRI), although imaging for OP is not specific. Biopsy is needed as a gold standard method especially for patients with recurrent or persistent condition.⁶ Some of

the differential diagnoses are Orbital cellulitis, Thyroid Associated Orbitopathy, Lymphangioma, Grave's Disease and Ruptured Dermoid Cyst.⁹ Another systemic disease which could be related with OP are Infection, Giant Cell Arthritis and Rheumatoid Arthritis.^{10,11}

Up to this day, the main treatment for OP is systemic corticosteroid. Oral corticosteroid may be started with initial dose of 1mg/kg of prednisone and tapered off for the next few weeks. Previous study reported that 27 patients were treated with oral corticosteroid, 21 of 27 patients were responded to the treatment, meanwhile 11 of 21 patients developed recurrence. Therefore, 10 of 27 patients treated with corticosteroid were controlled.⁴

Other study in Siriraj Hospital, Bangkok, also showed the improvement of 40 patients of total 49 patients who treated with oral corticosteroid.⁵ The potential of visual damage makes OP is something that should not be overlooked by ophthalmologists. We reported a case of OP and its management in clinical practices at a secondary hospital in peripheral area. According to previous studies, the gold standard of OP is with biopsy. However, due to the limitations at secondary hospital especially in peripheral area, the choices of additional examinations are limited. Therefore, the purpose of this paper is to help ophthalmologist to establish the diagnosis and treatment of OP especially in limited facilities hospital.

CASE PRESENTATION

A 47 year-old woman presented with the chief complaint swollen of the lower eyelid on the left eye (LE). She also had red eye, and pain on the LE. This conditions had begun 8 months ago before admission. She has visited two others hospital for the same condition, and diagnosed with orbital cellulitis, however the symptoms persisted until now. She had no complaint of fever, cough, myalgia, palpitation, excessive sweating or any

other complaint. She only had history of hypertension and secondary glaucoma on the LE. Meanwhile, the right eye (RE) remains normal and no history of trauma for both eyes. Ophthalmologic findings of RE were normal, while the LE has proptosis +4mm - +5mm, 1/300 of Visual Acuity (VA), Intraocular Pressure (IOP) N+3 through palpation, with a positive result of relative afferent pupillary defect (RAPD). Ocular motility was limited to 15 degree to latero-inferior direction. Slit lamp examination showed a chemosis and conjunctival injection, with corneal and anterior segment remains normal. Optic disc edema was found in funduscopy examination and no preauricular lymphadenopathy. Vital signs and laboratory results within normal limits.

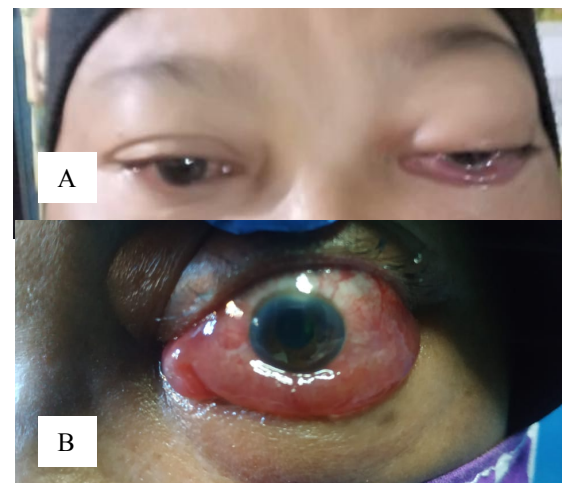


Fig 1. (A) (B) Patient's portrait in the first visit

Orbital CT Scan with and without contrast were performed, and showed a hypertrophy of ocular muscles and proptosis on LE with soft tissue swelling. Patient then was diagnosed with orbital pseudotumor (idiopathic orbital inflammation) and immediately prescribed with methylprednisolone with a dosage of 1 gr/day, divided into four daily increments. After discharge, patient was given methylprednisolone 16 mg 4x1 and tapered off after 1 weeks. There were a gradual improvement in protrusion, motility and visual acuity at two months follow up.

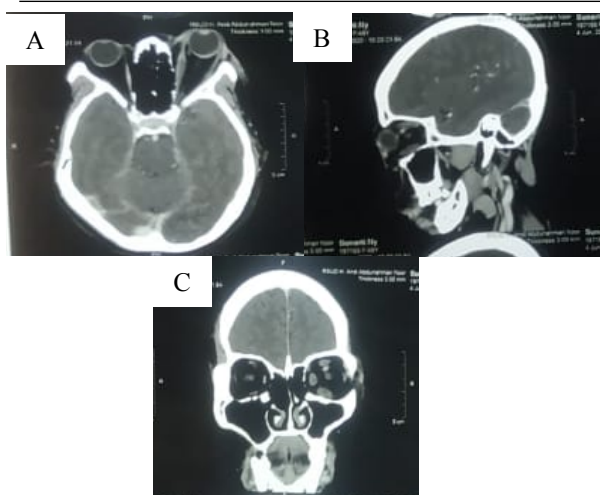


Fig 2. (A) (B) (C) Orbital CT Scan, Coronal, Axial, and Sagittal view

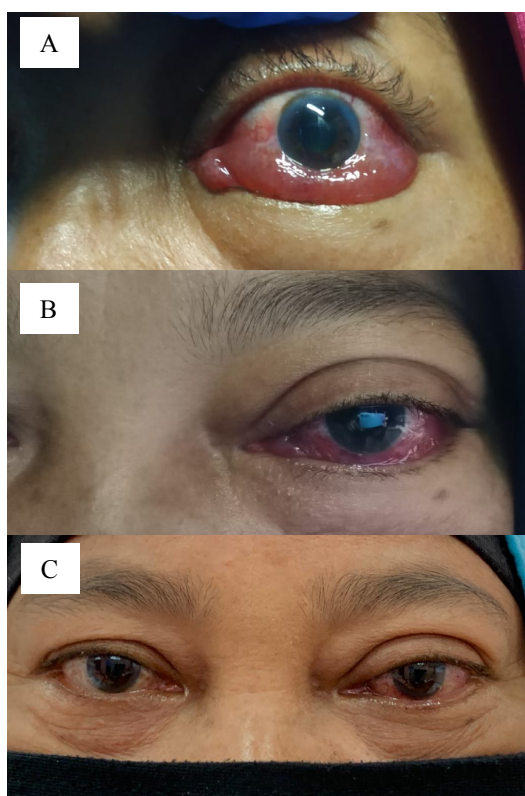


Fig 3. (A) Portrait of patient after 2 days of treatment. (B) Patient at 5th days of treatment (before discharge). (C) Clinical improvement at 2 months follow up.

DISCUSSION

OP is a non-inflammatory, non-neoplastic, non-specific process in the orbit and accounts approximately for 8-10% of all orbital tumors and the third most common cause of ophthalmologic tumors.^{2,7,10} Another study reported that the incidence of orbital tumor was 6% to 16%.^{12,13} OP is more common in people

with age ranging from 30 to 60 years old, especially in middle aged females.^{7,14} Recent study in Iran showed that the mean age of OP patients was 41 years old.¹³ In this case report, the patient is female, 47 years old and had the symptoms for 8 months with persistent symptoms, which appears to be a chronic and recurrence conditions, whereas in previous literatures stated that OP usually occurs in acute, sub-acute and occasionally chronic progression.^{6,14} Our patient had typical symptoms of OP, begin with swollen, red and pain on the LE. Jacobs and Galetta et al, stated that uncommon clinical sign of OP was decreased in visual acuity or visual loss, which is similar in our case with decreased in VA to 1/300.⁸ Orbital pseudotumor could mimics other diseases, therefore the differential diagnosis are varies, including Orbital Cellulitis, Thyroid Associated Orbitopathy (TAO), Grave's disease, Lymphangioma, Retrobulbar abscess, and Primary or Metastatic tumors.¹⁵ In extraordinary presentations, OP oftenly misdiagnosed with angioedema and temporomandibular joint dysfunction.^{15,16} However, in the present case there were no signs and symptoms of TAO, Grave's disease and Lymphangioma.⁸ Spindle et al, reported that OP usually occurs unilateral (in 88.2% patients), which is also in line with the present case.¹² Several studies reported that bilateral OP was more common in other inflammatory disease, like TAO and sarcoidosis, therefore it should be ruled out by a rheumatologic workup.^{7,8,17} Radiologic examination should be conducted to established the diagnosis. Orbital CT scan showed the thickening in extraocular muscle, along with proptosis and soft tissue swelling of the LE. Unfortunately, CT scan is not reliable enough to differentiated from other potential casues.^{17,18} In addition, the present case had recurrence symptoms for 8 months, therefore not only CT scan was needed, but also a biopsy was indicated. Biopsy of OP is necessary for patient with persistent or recurrence symptoms, except

the procedure would associated with significant risk of vision.^{6,14} In condition of diagnostic uncertainty, resistance and recurrence in steroid, biopsy is also suggested.¹⁰ Histologically, lymphocytes, plasma cells, neutrophils, macrophages and eosinophils may present in the biopsy results of OP.⁶ Unfortunately, due to the limited facilities at secondary hospital, biopsy could not be performed, therefore the diagnose of OP were establish by clinical presentations and CT scan. There are several option for treatment of OP. Radiotherapy is commonly used as the first line therapy for patients whom corticosteroids are contra indicated and as the second line for patients that has incomplete responds with corticosteroids.^{19,20} According to several studies, the success rate varies from 50% to 75%, with low dose radiation therapy at 1,500 – 2,000 centigray unit.^{20,21} Another studies reported that immunosuppressant agent have shown efficacy in the cases of OP²²⁻²⁴ Zacharopoulos et al, reported that there was a dramatic improvement after 6 weeks of Cyclosporin-A 4mg/kg per day.²² Smith and Rosenbaum treated seven patients of OP with methotrexate as a steroid-sparing drug with initial dose 7.5 mg/kg orally and four of seven patients had a better improvement.²⁵ Systemic corticosteroid is the main treatment of OP. Over 71% of patients had an improvement at 24 – 48 hours of corticosteroid. Oral corticosteroid may be started with initial dose at 1mg/kg of prednisone and when improvement is noted, dosage should be continued with a slow tapering over week to months, depends on each individual's response.^{7,8,18,26} Side effects of corticosteroid are, insomnia, mood swings, hypoglycemia, weight gain, hypertension and cushingoid faces.⁹ In line with our patient, who immediately prescribed with high dose of methylprednisolone 1 gr/day for 5 days and then tapered off after 1 week. Before discharge, patient had been informed about the side effect of corticosteroid, another treatment option if

there is no respond in corticosteroid and possibilities of differential diagnoses. After two months of follow up the patient showed dramatic improvements with no sign of side effects of corticosteroid. The authors certify that the patient had obtained all appropriate patient consent forms. Patient have given her consent for their images and other clinical information to be reported in the journal. The patient understand that her names and initials will not be published and due efforts will be made to conceal their identity.

CONCLUSION

Orbital pseudotumor is a condition that mimics a variety of diseases. Patients with OP needs complete history taking, physical examination and additional examination. In this case, the patient was admitted to secondary hospital in peripheral are with limited facilities, where MRI or biopsy could not be performed. Therefore the diagnosis was made through clinical presentation and CT Scan. A thorough history taking, comprehensive examination and choices of appropriate additional examination are needed in order to establish the diagnosis and management of OP to prevent further visual damage or dysfunction and gave an improvement for the symptoms.

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